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Sweet's Syndrome in Chronic Phase of Chronic Myeloid Leukaemia

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ACUTE FEBRILE NEUTROPHILIC DERMATOSIS, or Sweet's syndrome, is characterised by fever, neutrophilia and multiple, raised, erythematous, painful cutaneous plaques [1]. Neutrophilic infiltrate is found in skin lesions. The disease responds promptly to steroid therapy.

Sweet's syndrome is frequently associated with malignancies [2], meriting inclusion amongst paraneoplastic conditions. Several haematological malignancies have been associated with it [2]. However, only 3 cases have been reported in chronic myeloid leukaemia (CML): 2 in blast crisis [3, 4] and 1 in the accelerated phase of the disease [5].

Our patient, a 50-year-old man, was admitted in February 1986 with a temperature of 38°C and myalgias and painful, erythematous skin infiltrates on the trunk and arms. Skin lesions had first appeared a year earlier, and had been reappearing since in crops lasting 14 days, accompanied by fever. On admission, the patient had mild macrocytic anaemia (haemoglobin 125 g/l, mean corpuscular volume 101 fl), leukocytosis (white blood cells $23 \times 10^9/l$, with 2% promyelocytes, 5% myelocytes, 2% metamyelocytes, 2% bands, 69% neutrophils, 2% basophils, 3% monocytes and 15% lymphocytes), and a normal platelet count ($160 \times 10^9/l$). The leucocyte alkaline phosphatase score was nil. CML was confirmed by cytogenetic studies that showed the Ph chromosome in 50% of mitoses (46,XY/46,XY,Ph).

During one of the eruptions, a skin biopsy was done which revealed dense, perivascular infiltration of the dermis with neutrophils, lymphocytes and occasional histiocytes, without signs of vasculitis: findings typical of Sweet's syndrome. Therapy with oral corticosteroids led to disappearance of skin lesions and a decrease in their frequency.

The patient is presently, more than 5 years after the diagnosis of CML, in accelerated phase of the disease, managed with hydroxyurea. He is maintained on low-dose steroid therapy and has occasional skin eruptions, requiring the addition of parenteral betamethasone.

Sweet's syndrome is an established paraneoplastic condition, but it may also precede malignancy [2], as it did in our patient. It has rarely been associated with CML; moreover, all hitherto reported cases were diagnosed in advanced phases of the disease. Our case thus seems to be the first report of Sweet's syndrome in the chronic phase of CML.

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Family History of Oesophageal Cancer in Shanxi Province, China

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WE HAVE examined risk factors in a group of patients newly diagnosed with oesophageal cancer in Shanxi Province, China, an area where rates of this disease are among the highest in the world.

Interview and medical record information was collected on all 217 newly diagnosed cases seen at the Shanxi Tumor Hospital in Taiyuan during 4 months in 1989. A control group of 49 inpatients without cancer from the Shanxi Medical College No. 2 in Taiyuan was also interviewed.

Among cases, the median age at diagnosis was 56 years. 70% of cases were male, 59% were current smokers, 34% reported use of traditional Chinese alcoholic products, 50% consumed pickled vegetables and 20% reported a positive family history of oesophageal cancer. In contrast, only 1 control (2%) reported a positive family history of oesophageal cancer. The odds ratio (OR) for oesophageal cancer in subjects with a positive compared with those with a negative family history was 12.4 (95% CI 1.7-92.5). After adjustment for age and occupation, this OR was 7.9 (1.0-60.5).

This preliminary evaluation indicates that both established (i.e. tobacco and alcohol) and suspected (i.e. pickled vegetable consumption) risk factors for oesophageal cancer are common exposures in this high-risk region. Of particular interest is the potential role of genetic factors in the aetiology of oesophageal cancer in this area of China, where poor transportation and lack of access to the outside world may contribute to a peculiar genetic predisposition to this disease. We are exploring this possibility further, beginning with confirmation of familial aggregation of this disease.

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